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**Abstract:** Background Several aetiologies account for exercise intolerance, with cardiac sarcoidosis (CS) constituting a rare cause thereof. The pathogenesis of CS is still unresolved and its diagnosis still difficult to establish, in the absence of any extracardiac manifestations in particular. Case summary A 49-year-old amateur athlete presented with exercise intolerance during running over a 3-week period. Coronary artery and structural lung disease were excluded by coronary angiography and computer tomography. The symptoms could be reproduced during spiroergometry during which an exercise-induced high-degree atrioventricular (AV) block was documented. During electrocardiographic monitoring, a 2:1 AV block was observed. Different imaging modalities showed inferobasal septal inflammation and fibrosis. Transthoracic and transoesophageal echocardiography-guided endomyocardial biopsies were inconclusive and only subsequent epicardial biopsy performed by transdiaphragmatic minimally invasive surgery lead to the histological diagnosis of non-caseating granuloma, confirming CS. The patient was treated with high-dose steroids 1 week after implantation of a primary prevention dual-chamber implantable cardioverter-defibrillator (ICD). While tapering steroids, recurrence of myocardial inflammation occurred. However, no tachytherapies and <0.1% right ventricular pacing were needed after 2 years of follow-up. Discussion Differential diagnoses were either an infiltrative disease, a tumour, or an infectious disease. Due to the different treatment options, we had to establish definite diagnosis by myocardial biopsy. Retrospectively, the implantation of the ICD can be discussed. However, cardiac magnetic resonance imaging showed fibrosis which is usually irreversible and substrate for potentially lethal ventricular arrhythmia. Confirming the diagnosis of isolated CS is challenging. Long-term management should be guided individually based on clinical and imaging findings.

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# Exercise intolerance — from spiroergometry to transdiaphragmatic myocardial punch biopsy: a case report of isolated cardiac sarcoidosis

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## Background

Several aetiologies account for exercise intolerance, with cardiac sarcoidosis (CS) constituting a rare cause thereof. The pathogenesis of CS is still unresolved and its diagnosis still difficult to establish, in the absence of any extracardiac manifestations in particular.

## Case summary

A 49-year-old amateur athlete presented with exercise intolerance during running over a 3-week period. Coronary artery and structural lung disease were excluded by coronary angiography and computer tomography. The symptoms could be reproduced during spiroergometry during which an exercise-induced high-degree atrioventricular (AV) block was documented. During electrocardiographic monitoring, a 2:1 AV block was observed. Different imaging modalities showed inferobasal septal inflammation and fibrosis. Transthoracic and transoesophageal echocardiography-guided endomyocardial biopsies were inconclusive and only subsequent epicardial biopsy performed by transdiaphragmatic minimally invasive surgery lead to the histological diagnosis of non-caseating granuloma, confirming CS. The patient was treated with high-dose steroids 1 week after implantation of a primary prevention dual-chamber implantable cardioverter-defibrillator (ICD). While tapering steroids, recurrence of myocardial inflammation occurred. However, no tachytherapies and <0.1% right ventricular pacing were needed after 2 years of follow-up.

## Discussion

Differential diagnoses were either an infiltrative disease, a tumour, or an infectious disease. Due to the different treatment options, we had to establish definite diagnosis by myocardial biopsy. Retrospectively, the implantation of the ICD can be discussed. However, cardiac magnetic resonance imaging showed fibrosis which is usually irreversible and substrate for potentially lethal ventricular arrhythmia. Confirming the diagnosis of isolated CS is challenging. Long-term management should be guided individually based on clinical and imaging findings.

## Keywords

Exercise testing • AV block • Imaging • Biopsy • Sarcoidosis • ICD implantation • Case report

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## Learning points

- Spiroergometry is a useful tool for work-up of exercise intolerance.
- Diagnosis of isolated cardiac sarcoidosis is difficult. Cardiac biopsy is crucial for definite diagnosis in the absence of any extracardiac manifestations.
- If endomyocardial biopsies remain unspecific, a surgical epicardial approach can be helpful—especially when pathology is involving more the epicardial than endocardial layers.
- Cardiac sarcoidosis is associated with sudden cardiac death due to both brady- and tachyarrhythmias.
- Regular follow-up visits are needed for steroid-dose adjustment based on clinical and imaging findings.

## Introduction

Exercise intolerance is a common symptom in sports cardiology with a broad differential diagnosis, ranging from common coronary artery disease to rare findings, e.g. cardiac sarcoidosis (CS) among several other rare aetiologies. Pathogenesis of CS remains unresolved and diagnosis often challenging, in the absence of extracardiac manifestations in particular.<sup>1</sup> Up to 5% of patients with sarcoidosis present with clinically manifest and another 20% of patients with asymptomatic, clinically silent cardiac involvement.<sup>2</sup> The clinical features of CS, especially advanced conduction system disease and/or heart failure are dependent on the location, extent, and duration of the disease.<sup>3</sup>

## Case presentation

A 49-year-old amateur athlete presented with exercise intolerance (by means exertional dyspnoea and dizziness) during running for 3 weeks. The patient has not had a syncopal episode or family history of sudden cardiac death (SCD). He did not take any prescription or non-prescription drugs. The travel history revealed several trips to Iran in the past years. His medical history revealed an inguinal bullous and crusty cutaneous lesion of 4 cm × 5 cm 3 months prior to initial presentation that had been treated by a physician in Italy during the summer holidays with topical steroid application for a few days. No recurrence occurred.

At initial presentation at our hospital, vital signs and clinical examination were unremarkable (heart rate at 61 b.p.m., blood pressure

## Timeline

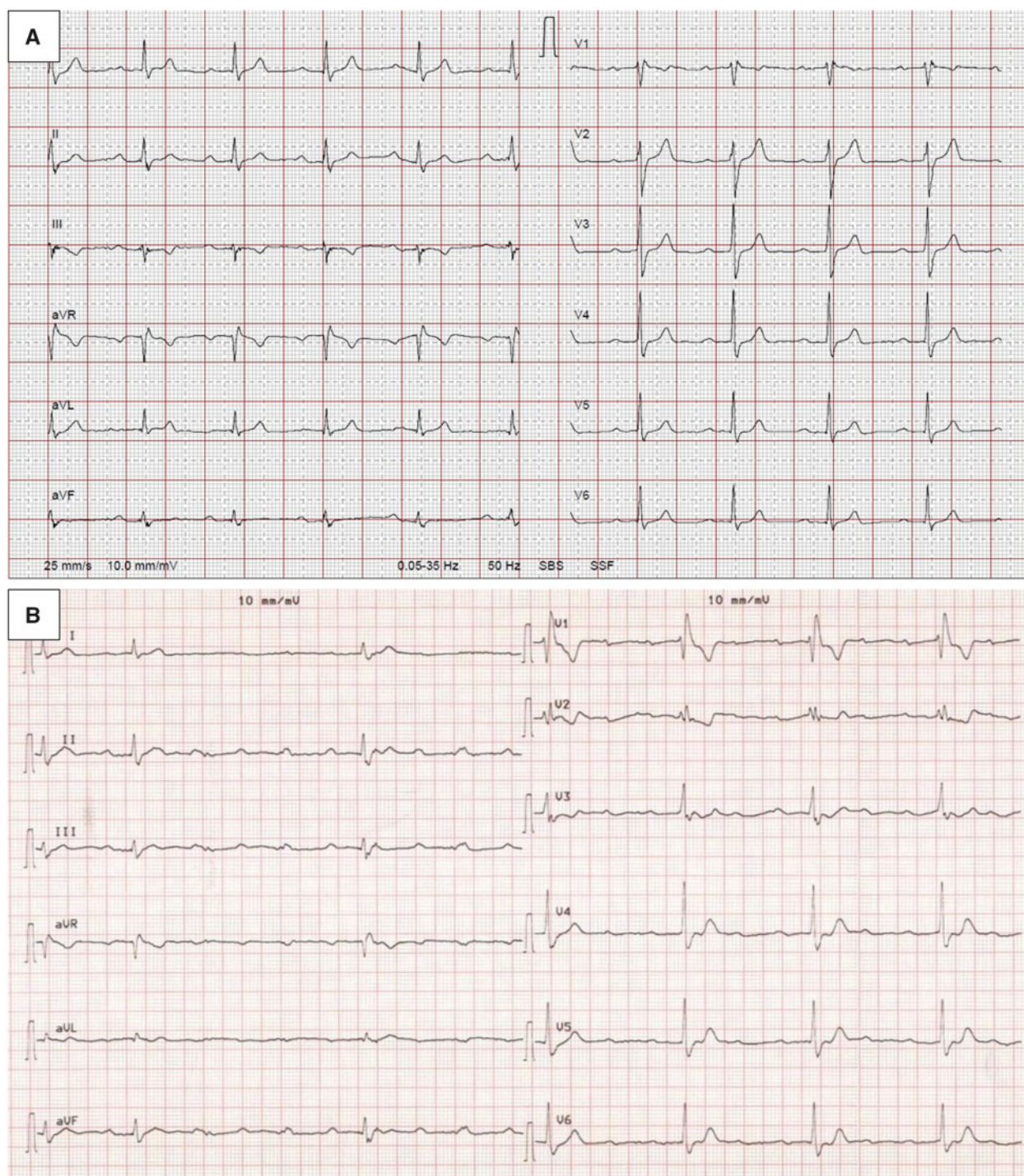
### Work-up

Days	
0	Electrocardiogram (ECG), echocardiography, coronary angiography, chest computer tomography scan, Holter-ECG
2	Cardiac magnetic resonance imaging: inferobasal mass
5	Spiroergometry: high-degree atrioventricular block
8	<sup>18</sup> F-fluorodeoxyglucose-positron emission tomography (FDG-PET): focal septal inflammation, no extracardiac manifestations
14	Transthoracic echocardiography-guided endomyocardial biopsy (EMB)
21	Deep vein thrombosis → anticoagulation
22	Transoesophageal echocardiography-guided EMB
33	Transdiaphragmatic cardiac punch biopsy
41	Dual-chamber implantable cardioverter-defibrillator (ICD) implantation

### Follow-up

Months after ICD implantation		Steroid dose (mg) qd
0	Initiating steroids 0.8 mg/kg	60
1	Patient asymptomatic, steroid tapering over months	50
2	Stop anticoagulation	50
6	FDG-PET: no inflammation	35
10	FDG-PET: recurrence of inflammation	17.5 → 40
13	Azathioprine 50 mg qd	20
15	Azathioprine stop (nausea)	17.5
20	FDG-PET: no inflammation	12.5
25	FDG-PET: mild inflammation	10 → 20
26	Methotrexate 10 mg qwk	15

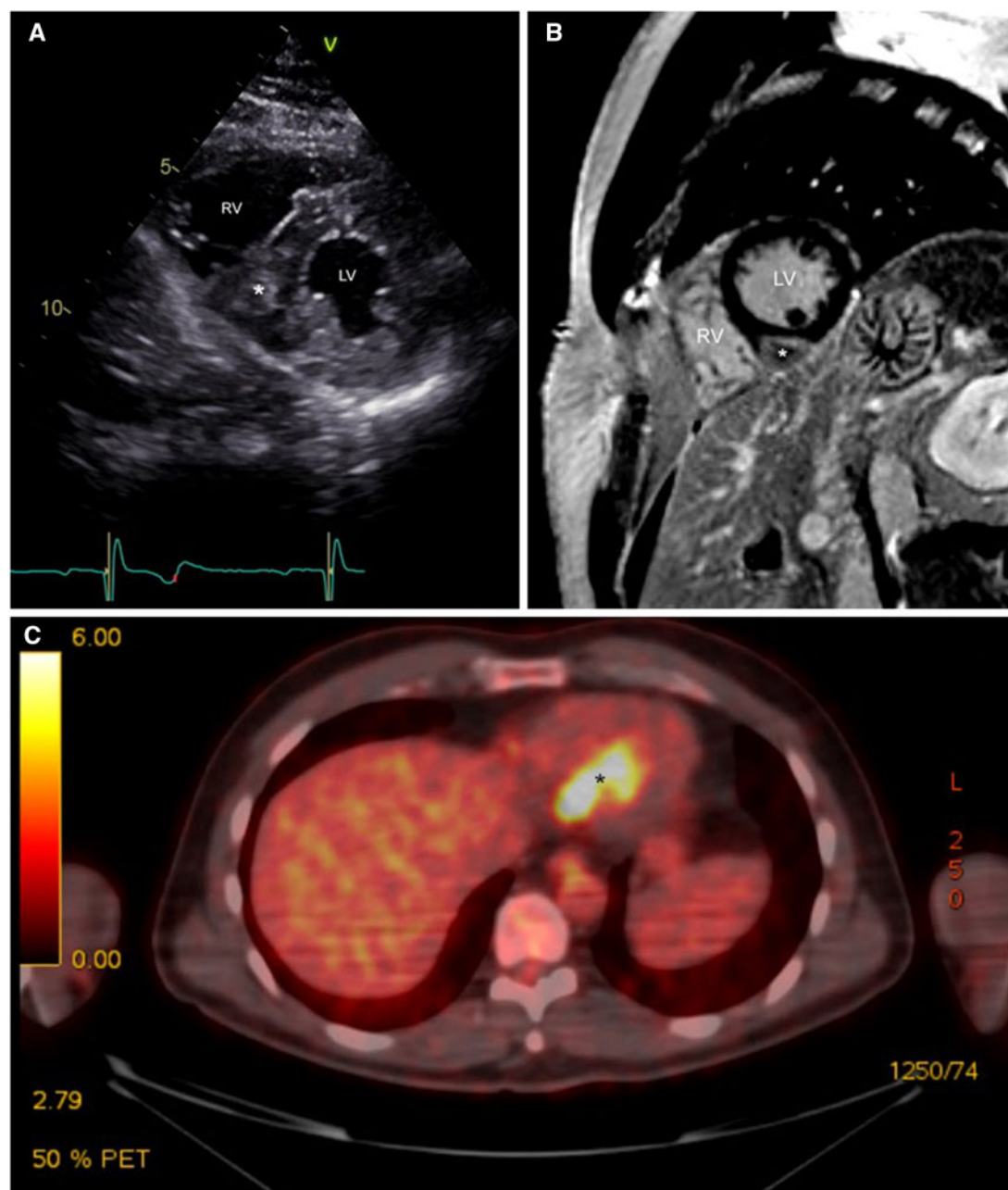




**Figure 1** (A) Initial resting 12-lead electrocardiogram. Incomplete right bundle branch block and atrioventricular block I°. Calibration 10 mm/mV and speed 25 mm/s. (B) 12-lead electrocardiogram. High-degree atrioventricular block during spiroergometry at 90 W. Calibration 10 mm/mV and speed 25 mm/s.

122/77 mmHg, temperature 36.5°C, normal cardiac and pulmonary examination). D-dimers and inflammation markers were negative [leucocytes 4.89 g/L (normal 3–9.6 g/L), C-reactive protein 0.8 mg/L (normal <5 mg/L)].

Resting 12-lead electrocardiogram showed an incomplete right bundle branch block with atrioventricular (AV) block I° (Figure 1A). Rhythm monitoring showed intermittent 2:1 AV block. Performing a spiroergometry, we were able to reproduce the symptoms while



**Figure 2** (A) Transthoracic echocardiography, short axis. Intracardiac mass located in the inferobasal septum. (B) Cardiac magnetic resonance imaging, short axis. Late gadolinium enhancement in the inferobasal septum. (C)  $^{18}\text{F}$ -fluorodeoxyglucose-positron emission tomography. Intense uptake in the inferobasal septal region. LV, left ventricle; PET, positron emission tomography; RV, right ventricle. \*Intracardiac mass.

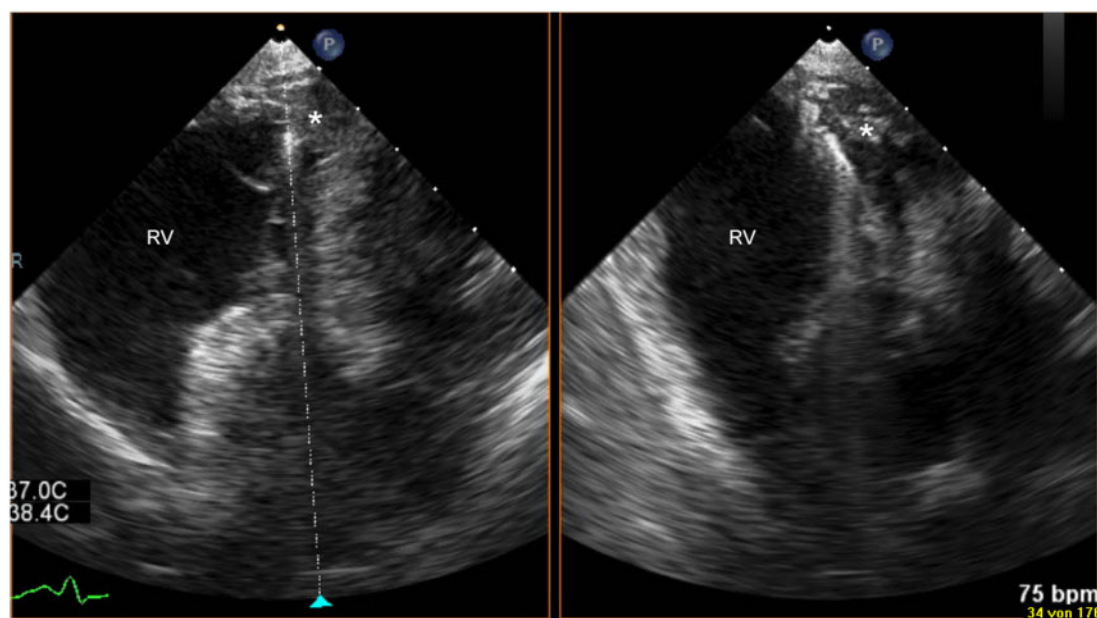
documenting an exercise-induced high-degree AV block (Figure 1B). Initial transthoracic echocardiography raised questionable wall motion abnormalities and in retrospect, a mass in the inferobasal septum (Figure 2A). Left ventricular ejection fraction was normal. Subsequent coronary angiography excluded coronary artery disease and computer tomography of the chest did not reveal any structural pulmonary disease. Cardiac magnetic resonance imaging (CMR) showed an intracardiac mass involving the inferobasal septum describing a formation of 5 cm × 1 cm × 1 cm. Late gadolinium enhancement (LGE)

was positive at this particular site (Figure 2B). However, no active perfusion of this particular site was seen in coronary angiography nor CMR.

$^{18}\text{F}$ -fluorodeoxyglucose-positron emission tomography (FDG-PET) documented an intense uptake of this mass, indicating an inflammatory process (Figure 2C).

Clinical examination, abdominal ultrasound, and whole-body FDG-PET did not demonstrate any extracardiac focus or lesion. In addition, inflammation markers were negative (sIL-2 receptor,





**Figure 3** Transoesophageal echocardiography. Guiding of endomyocardial biopsy in two planes. RV, right ventricle. \*Intracardiac mass.

antinuclear antibodies, anti-neutrophil cytoplasmic antibodies), serum calcium and angiotensin-converting enzyme were normal [2.38 mmol/L (normal 2.09–2.54 mmol/L), 16.5 U/L (normal 12–68 U/L)]. Microbiology did not reveal any abnormalities: blood cultures for aerobic and anaerobic bacteria and mycobacteria were negative. *Brucella* species, *Bartonella henselae*, *Treponema pallidum*, *Borrelia burgdorferi*, and *Coxiella burnetii* antibodies as well as the result of the HIV test were all negative. Travel history to Iran raised pre-test probability for mycobacterial infection. However, polymerase chain reaction for tuberculosis and blood cultures for mycobacteria remained negative. Dermatologist-performed full-body skin examination revealed only unremarkable cutaneous lesions that were considered as non-specific and not typical for sarcoidosis.

So far, blood tests did not reveal any further abnormalities.

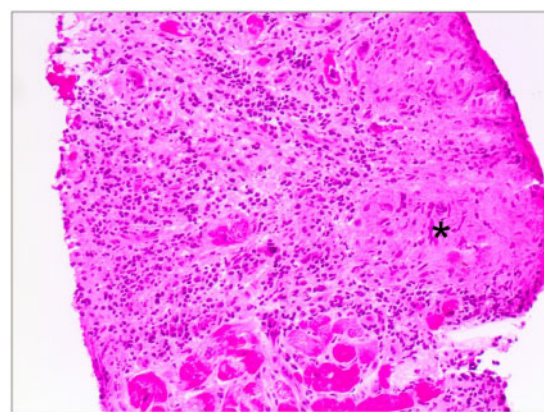
Due to lack of extracardiac manifestations, we attempted to confirm diagnosis by endomyocardial biopsy (EMB). Despite optimal transthoracic and transoesophageal echocardiographic guiding (Figure 3), however, all samples remained non-specific.

Moreover, a deep vein thrombosis (DVT) occurred despite adequate thrombosis prophylaxis during hospitalization, underlining the active (inflammatory) process. After initial anticoagulation with heparin, edoxaban (60 mg qd) was started for treatment of DVT.

Given its location in the inferobasal septum, and as the pathology was involving more the epicardial than endocardial layer, we performed an epicardial punch biopsy on the beating heart via a minimally invasive transdiaphragmatic approach thus avoiding open-heart surgery.

Histological examination of this punch biopsy demonstrated non-caseating granuloma (Figure 4), highly suggestive for CS.

In summary, after thorough exclusion of any extracardiac manifestations, histological results confirmed criteria for an isolated CS.<sup>2</sup>



**Figure 4** Histological sample from the inferoseptal right ventricle (haematoxylin and eosin stain). \*Non-caseating granuloma.

Due to ventricular tachyarrhythmias and/or AV conduction block, SCD in CS is reported in up to 30–65%. Due to the presence of LGE on CMR—implicating potential irreversible fibrosis—and intermittent high-degree AV block, we felt that there was the need for a dual-chamber device. Hence, we decided to insert a transvenous dual-chamber implantable cardioverter-defibrillator (ICD) for primary prevention of SCD and brady pacing. To facilitate wound healing and reducing the risk of wound infection after ICD implantation, we started high-dose steroids (prednisone 0.8 mg/kg qd) 1 week after implantation.

After establishing steroids, PR-interval shortened from 224 to 150 ms. Four weeks later, at the first follow-up visit, the patient was already asymptomatic during exercise with 1:1 AV conduction during effort. Steroids were tapered over time (~10 mg less every 1–2 months). To avoid or diminish side effects of long-term immunosuppression with steroids, pneumocystis pneumonia prophylaxis with trimethoprim-sulfamethoxazole, gastric protection with pantoprazole, and osteoporosis prophylaxis with calcium, vitamin D, and alendronic acid were established. Due to recurrence of myocardial inflammation (FDG-PET) after 10 months, we increased the steroid-dose and initiated additive immunosuppressive therapy with azathioprine 3 months later to diminish side effects of long-term steroid intake. Because of nausea, this approach had to be abandoned. As there were no signs of recurrence of myocardial inflammation (FDG-PET) steroids were tapered again. After recurrence of mild inflammation, methotrexate was initiated in a dose of 10 mg per week and subsequently increased.

During last follow-up, 2 years after ICD implantation, no tachytherapies and <0.1% right ventricular (RV) pacing by the ICD were necessary.

## Discussion

Differential diagnoses of the intracardiac inflammation process, documented by multi-modality imaging, were an infiltrative disease, a tumour or an infectious disease (travel history to Iran and Italy). Theoretically, the above-mentioned skin lesions could have been an earlier, cutaneous manifestation of CS. Subsequent full-body skin evaluations by a dermatologist in our clinic, however, did not detect any signs of cutaneous sarcoidosis.

Malignant and infectious diseases could not be completely ruled out, and histopathological evaluation is required for the definite diagnosis of isolated CS.<sup>2,4</sup> As the FDG-PET and CMR results were suggestive of CS, an EMB was performed. While invasive electroanatomical voltage mapping by electrophysiologists would provide optimal guiding for an EMB, the inferobasal septal lesion had been visible on echocardiography. Despite meticulous guiding by echocardiography, the biopsy specimens obtained by EMB of the right ventricle did not show any CS. Therefore, we proceeded with an epicardial punch biopsy on the beating heart via a minimally invasive transdiaphragmatic approach that finally confirmed the diagnosis of CS. Indeed, histopathological evaluation demonstrated that the sarcoid lesions involved more the epicardial than endocardial layers. Of note, sensitivity of EMB in CS is reported to be as low as 20%.<sup>5</sup>

In view of the paucity of data to guide clinical care in patients with CS, current management recommendations in CS are still largely based on expert opinion. In particular, whether patients with advanced conduction system disease and preserved ventricular function should receive a bradypacemaker or an ICD is still matter of debate.<sup>2,6</sup> Results from observational studies, such as the MIDFIN Study Group (Myocardial Inflammatory Diseases in Finland)<sup>3</sup> as well as by Nordenswan et al.<sup>7</sup> add further support to the recent AHA and HRS Expert Consensus Statement recommendation supporting ICD implantation in CS patients with advanced conduction system disease.

Importantly, clinical equipoise exists, as ICD complications have been shown to be elevated in CS populations.<sup>7,8</sup> This notwithstanding we decided to implant a dual-chamber ICD for prevention of SCD due to brady- or tachyarrhythmia<sup>2,9</sup> as there was LGE observed on CMR, which may reflect cardiac fibrosis. Recurrence of inflammation raises probability of fibrotic remodelling. Usually, fibrosis is irreversible and implicates a substrate for potential fatal arrhythmia. However, shared decision-making with the informed patient is of utmost importance.

Of note, after 2 years of follow-up, the patient did not need any tachytherapies and RV pacing was <0.1%.

For optimal wound healing after ICD implantation, we waited with steroid treatment for 1 week. Currently, there exist no randomized studies about detailed steroid use, i.e. starting dose, duration, tapering, and other immunosuppressive regimen. Few studies of CS with conduction abnormalities underline the effect of steroids.<sup>10,11</sup> Based on our local expertise, we decided to start steroids with 0.8 mg/kg (60 mg qd) under the aspect of high-degree AV block and focal inflammation. Tapering of steroids and steroid-sparing agents should be achieved over the first months and years of follow-up to diminish side effects of long-term steroid intake. As the patient remained asymptomatic, we tried to guide tapering with repetitive cardiac FDG-PET imaging, which has to be weighed against potential long-term radiation side effects.

In summary, imaging was already suggestive of CS. Confirmation of the diagnosis of CS was subsequently obtained by cardiac biopsy. The treatment regimen is based on two pillars: suppression of inflammation with steroids and prevention of SCD due to brady- and tachyarrhythmias with an ICD. Regular follow-up visits are of utmost importance for adjustment of immunosuppression.

## Conclusion

Isolated CS is a rare, but not irrelevant differential diagnosis of exercise intolerance, especially along with conduction abnormalities during effort. Definite diagnosis is difficult to obtain but crucial for adequate therapy. Management of follow-up remains difficult due to individual disease progression and handling of immunosuppression.

## Lead author biography



Dr Michael Johannes Schindler is a Specialist in Cardiology, spec. Sports Medicine (SGSM), Department of Preventive Sports Medicine and Sports Cardiology, Klinikum rechts der Isar, Technical University, Munich. His residency was at the Heart Centre, University Hospital of Zurich, and Cantonal Hospital of Baden, Switzerland. He graduated from the University of Zurich. His research interest is in cardiac imaging of sports adaptation.



## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** Dr. BENUSSI reports personal fees from ATRICURE, INC., outside the submitted work. FR reports grant support for the ESC-HFA Postgraduate Course in Heart Failure from Novartis, Servier, Bayer, Abbott and Astra Zeneca and the VASCEND trial from Novartis (all payments directly to the University of Zurich). FR has been paid for the time spent as a committee member for clinical trials, advisory boards, other forms of consulting and lectures or presentations. These payments were made directly to the University of Zurich and no personal payments were received in relation to these trials or other activities since January 2018. Before 2018 FR reports grants and personal fees from SJM / Abbott, Servier, Novartis and Bayer, personal fees from Zoll, Astra Zeneca, Sanofi, Amgen, BMS, Pfizer, Fresenius, Vifor, Roche, Cardiorentis and Boehringer Ingelheim, other from Heartware and grants from Mars, outside the submitted work. Dr. Saguner reports educational grants from Abbott, Biosense Webster, Boston Scientific, BMS-Pfizer, Bayer Healthcare, Biotronik, Medtronic and holds shares from Gilead Sciences. Dr. Schindler reports personal fees from Amgen, outside the submitted work. Dr. Steffel reports personal fees from Amgen, personal fees from Astra Zeneca, grants and personal fees from Bayer Healthcare, personal fees from Boehringer-Ingelheim, grants and personal fees from Biosense Webster, grants and personal fees from Boston Scientific, personal fees from Bristol-Myers Squibb, grants and personal fees from Daiichi-Sankyo, grants and personal fees from Medtronic, personal fees from Novartis, personal fees from Pfizer, personal fees from Sanofi-Aventis, grants and personal fees from Abbott, personal fees from Zoll, other from CorXL, grants and personal fees from Biotronik, personal fees from Atricure, personal fees from Medscape, personal fees from WebMD, personal fees from Merck / MSD, personal fees from Berlin Chemie / Menarini, outside the submitted work. Dr. Steffel reports personal fees from Amgen, personal fees from Astra Zeneca, grants and personal fees from Bayer Healthcare, personal fees from Boehringer-Ingelheim, grants and personal fees from Biosense Webster, grants and personal fees from Boston Scientific, personal fees from Bristol-Myers Squibb, grants and personal fees from Daiichi-Sankyo, grants and personal fees from Medtronic, personal fees from Novartis, personal fees from Pfizer, personal fees

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